EDF Euroderm Excellence Course 2025

Workshop 2 (Part 1): Great results in vitiligo, lupus, dermatomyositis, morphea, Behçet's, vasculitis

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Dermatomyositis: Cutaneous manifestations

Characteristic

- Gottron's papules
- Gottron's sign
- Holster sign
- Heliotrope erurption (facial erythema)
- Shawn and V-sign
- Periungual changes (capillary etc)
- Mechanic's hands

Unspecific

- Calcinosis cutis
- Psoriasiform eruption in scalp
- Diffuse alopecia
- Hyperpigmentation
- Erythrodermia
- Pruritus
- Panniculitis, nodules.. etc
- etc

Dermatomyositis: extracutaneous symptoms and signs

- Muscle weakness: at presentation in 50-60%, often symmetrical and proximal, typically at the same time or after development of skin symptoms. Some have myalgia and muscle tenderness
  - *Creatine kinase (CK), lactate dehydrogenase (LDH), aldolase, AST/ALT. Myositis-specific/associated autoantibodies*
  - *Muscle strenght testing, Electromyography (EMG), MRI, muscle biopsy*
- Interstitial lung disease (ILD): 30-40% of DM patients; before, at the time or after DM rash. Different types, nonspecific interstitial pneumonia most common (NSIP). Dyspnea and nonproductive cough or asymptomatic. Chest auscultation clear or bibasilar cracles.
  - *ESR, Chest radiography, HRCT if necessary. Pulmonary function tests help to distinguish the cause of dyspnea and to assess the severity of respiratory impairment.*

Dermatomyositis: extracutaneous symptoms and signs

- **Polyarthritis**: joint pain an swelling (Erythrocyte sedimentation rate, ESR, may be elevated)
- **Cardiac involvement:** Myocarditis, increased risk for myocardial infarction. Subclinical symptoms frequent (conduction abrnormalities, arrhythmia), symptomatic cardiac disease (congestive heart failure) rare.
- **Esophageal involvement:** Dysphagia, regurgitation, aspiration
- **Malignancy-related:** Risk for overall malignancies increased (x5-7). Most commonly within 0-1 years of diagnosis. Increased risk in older and non-responsive DM patients, those with ulcerative skin lesions and specific myositis-specific autoantibodies.
  - Individual investigations (chest radiography, ultrasound/CT, colonoscopy, mammography, gynekological investigations, PET-CT..

Dermatomyositis: classification

- Classic dermatomyositis: simultaneous cutaneous and muscle involvement (muscle weakness and presence of myositis in diagnostic tests)
- Amyopathic dermatomyositis (dermatomyositis sine myositis): DM cutaneous symptoms (for at least 6 months) but no muscle weakness or laboratory or radiological signs of myositis. Obs immunosuppressive medication!
- Hypomyopathic dermatomyositis: DM cutaneous symptoms (for at least 6 months) but no muscle weakness but subclinical evidence of myositis in laboratory or radiological investigations.
- Postmyopathic dermatomyositis: Persistence of DM cutaneous symptoms after resolution of muscle disease (with immunosuppressive therapy)
- Juvenile dermatomyositis >< Adult dermatomyositis

Myositis-specific antibody-defined subgroups

- **Antisynthetase antibodies and syndrome**: Presence of an antibody directed against one or several tRNA synthetases (most often anti-Jo-1). Inflammatory myopathy, ILD, mechanic's hands, inflammatory arthritis, Raynaud phenomenon.
- **Anti-Mi2 antibody**: relatively acute onset with classical cutaneous findings, more severe muscle disease, may respond well to therapy. Antibodies correlate with disease severity. Lower risk of malignancy?
- **Anti-TIF-1gamma antibody**: Palmar hyperkeratotic papules, psoriasis-like lesions, hypopigmented and telangiectatic patches. Strongly associated with increased risk of cancer (ovarian cancer particularly).
- **Anti-SAE antibody**: classical cutaneous findings, high prevalence of dysphagia, cutaneous manifestatins often precede myopathy. May have increased risk of malignancy
- **Anti-NXP2 antibodies**: juvenile dermatomyositis and calcinosis, increased risk of malignancy

Mammen et al. Neuromuscul Disord 2020;30:70-92.

Anti-MDA5 antibody-positive dermatomyositis

- = Anti-melanoma differentiation-associated protein 5
- Typical skin findings: Skin ulceration often overlying on Gottron's papules/signs and fingers, painful palmar papules, oral ulcerations, non-scarring alopecia
- Rapidly progressing interstitial lung disease (40-100%)
- Joint pain and arthritis (small joints in hands and wrists), cardiac involvement?
- Hoarseness is common, but severe dysphagia rare
- Peripheral lymphopenia, elevated serum ferritin levels. Infections common complications
- No (or mild) muscle involvement
- Poor response to systemic glucocorticoids and conventional immunosuppressents
- The risk of malignancies low
- Poor prognosis

Mammen et al. Neuromuscul Disord 2020;30:70-92, Lu et al. Nat Rev Rheumatol;20:48-62.

Treatment of cutaneous dermatomyositis

- Photoprotection
- Topical therapy: corticosteroids, calcineurin inhibitors
- Systemic therapy:
  - 1) hydroxychloroquine and quinacrine (mild disease), methotrexate (more severe)
  - 2) IVIG, Mycophenolate mofetil
  - 3) Rituximab, azathioprine, JAK-inhibitors, cyclosporine, dapsone etc.
  - Myositis often treated with systemic corticosteroids

- Challenges: 1) cutaneous manifestations are often rather resistant to therapy 2) there is scarce evidence of therapies 3) extracutaneous (e.g. myositis) disease and disease severity influence treatment decisions

Chronic ulcers

Chronic ulcers

- Venous ulcers
- Arterial ulcers
- Mixed ulcers
- Pressure ulcers
- Diabetic foot ulcers

"Typical ulcers"

- Atypical ulcers
  - Diverse group of ulcers caused by malignancy, inflammation, systemic or skin diseases, medication, etc.

10-20% of all chronic ulcers

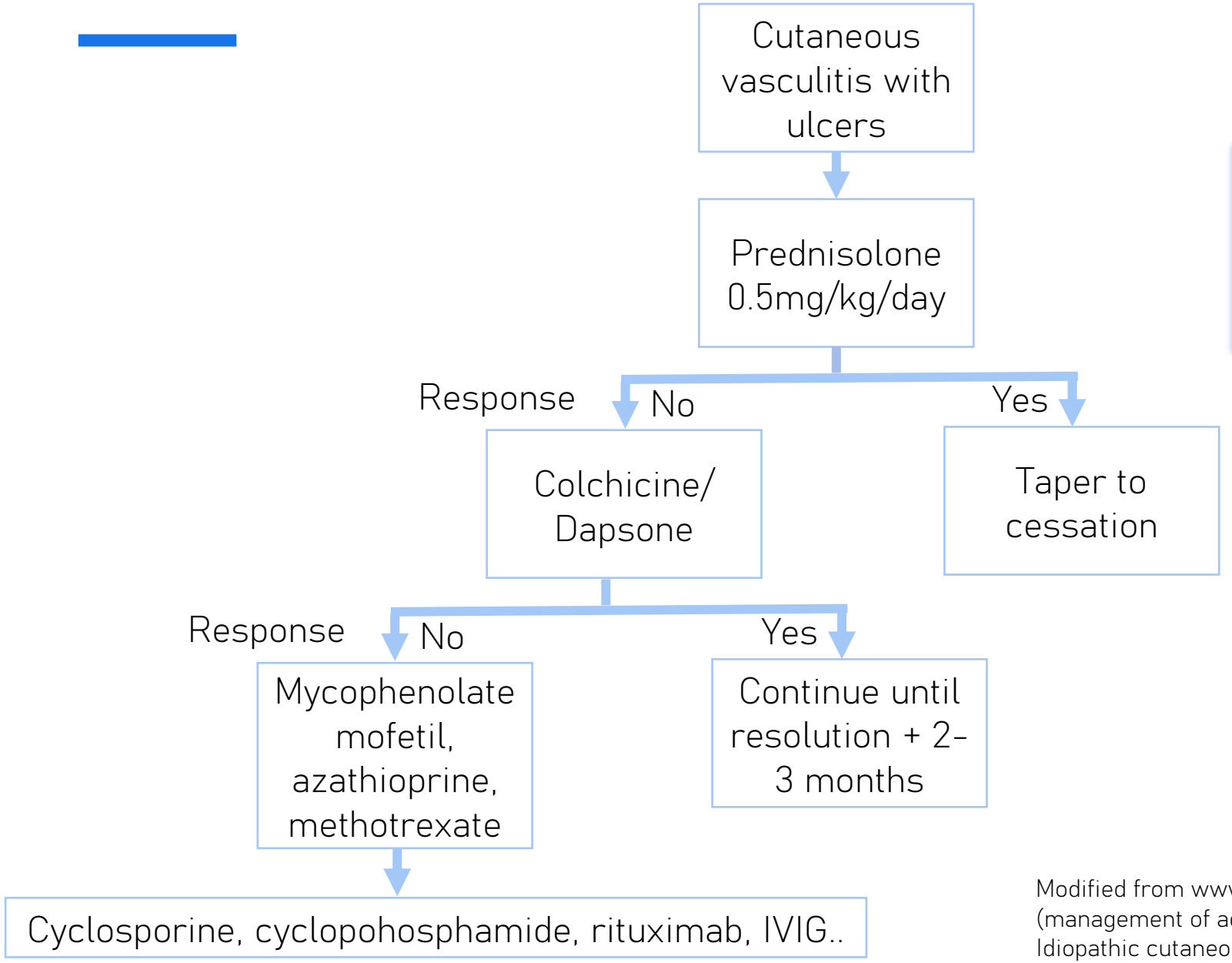
Isoherranen et al. J Wound Care 2019;28:S1-92.

Vasculitis ulcers: key issues

- 1) Most common atypical ulcer
- 2) App 5% of all chronic ulcers, incidence increasing
- 3) Typical clinical picture
  - Multiple, necrotic, painful ulcers with violaceous borders
  - Palpalable purpura/livedo racemosa in surrounding skin
- 5) Exclusion of systemic vasculitis and investigation of aetiology
  - *(e.g. thorough history + clinical examination, full blood count, renal and liver function tests, CRP, ESR, ANA-ab, ENA, ANCA-Ab, Pr3-Ab, MPO-Ab, complement factors, Rf, CCP-ab, cryoglobulins, Hepatitis B/C and HIV serology, Antiphospholipic and anticardiolipin antibodies, radiologic investigations, stool blood….)*

Körber et al. JDDG 2011;9:116-121.
Kimpimäki et al. Acta Derm Venereol 2017;97:653-4.
Isoherranen et al. J Wound Care 2019;28:S1-92.

Treatment of vasculitic ulcers



Medical treatment is based on
1) Specific diagnosis and aetiology
2) Severity of the disease
3) Patient's medical history

Modified from www.uptodate.com
(management of adults with
Idiopathic cutaneous small vessel vasculitis)

Is there a role for surgery in the management of vasculitic ulcers?

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ORIGINAL ARTICLE

IWJ WILEY

The role of split-thickness skin grafting in the treatment of vasculitic and pyoderma gangrenosum ulcers in a multidisciplinary wound centre

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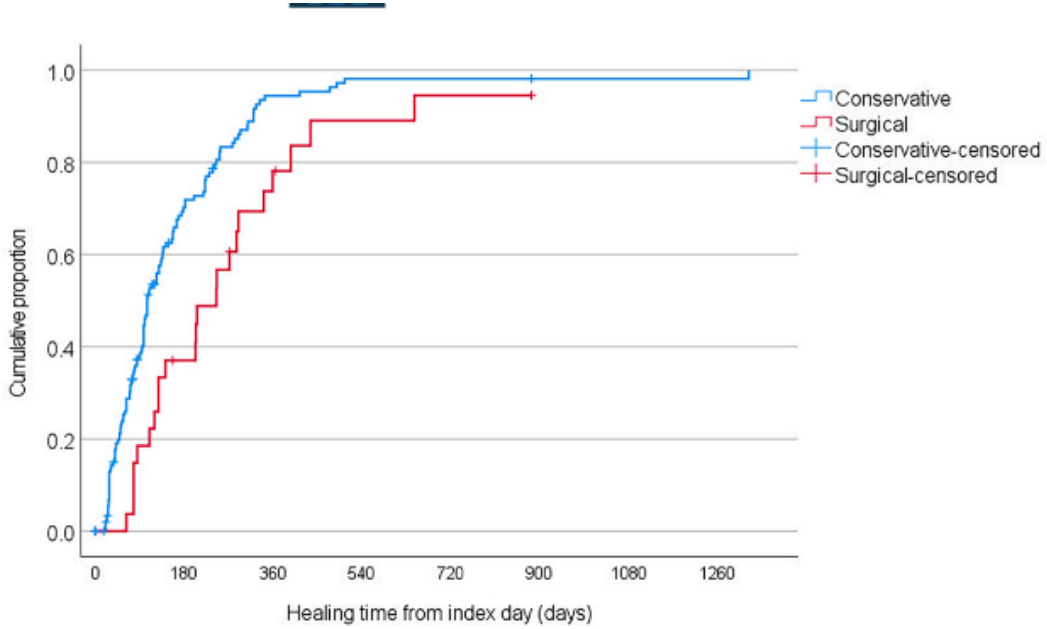
46 patients with vasculitic ulcers (and 107 Vus)
(34 patients with PG ulcers 74 PGUs)

- 9/46 (19.6%) of VU patients (25 out of 107 ulcers)
(2/34 (5.9%) of PG patients and 2 out of 74 ulcers)
were treated surgically
(large ulcer size, uncontrollable pain, delayed healing, recurring infections)



Surgically treated patients had more long-term illnesses, Vasculitis was more severe and ulcers larger than in those treated conservatively

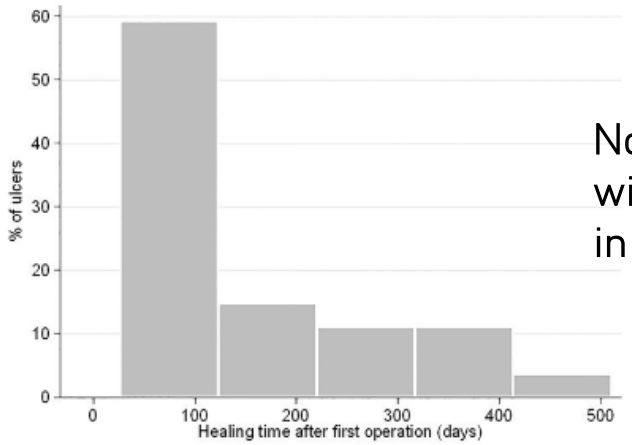
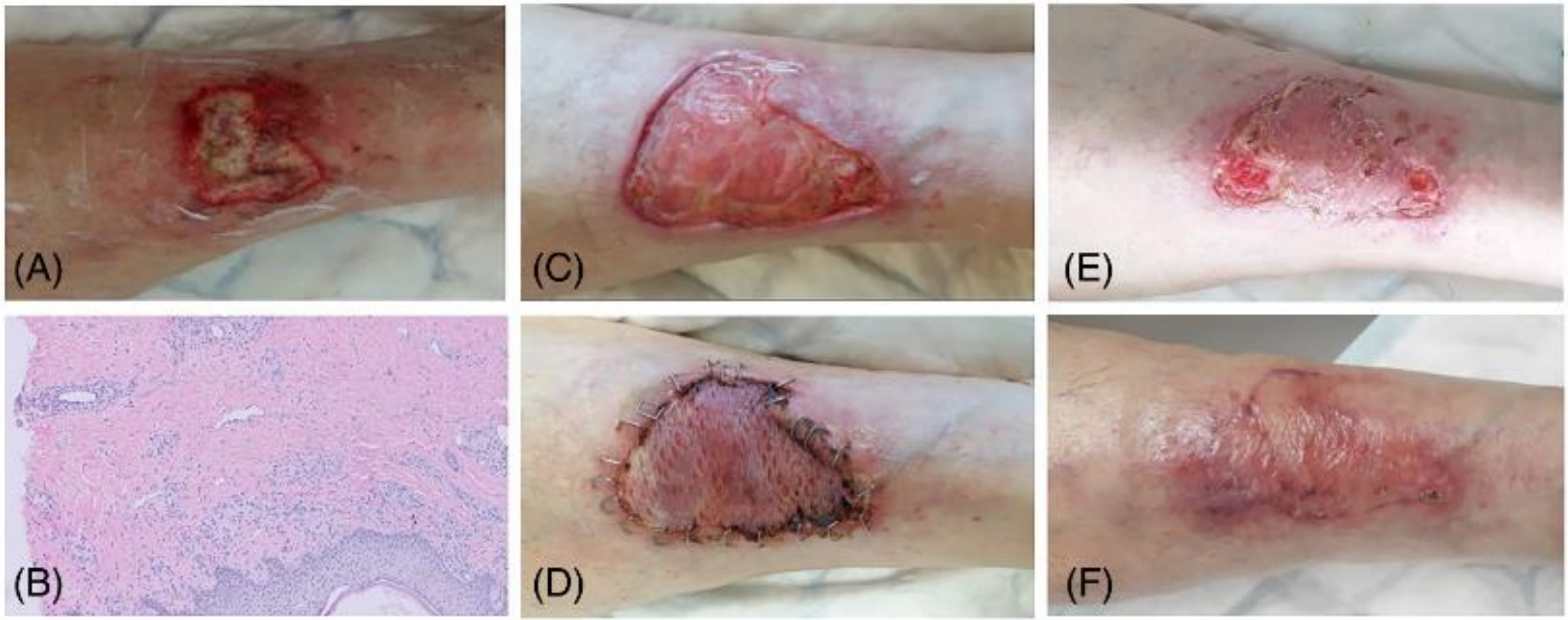


FIGURE 3 Healing time of surgically treated atypical ulcers ($n = 27$) after the first skin graft operation. Each gray bar represents proportion of ulcers that were healed in each timespan.

No pathergy related with surgery, no differences in ulcer recurrence or mortality



Suoniemi et al. Int Wound J 2024;21:e14943.

Vasculitic ulcers

- Diagnosis without delay
- Thorough examinations at diagnosis (patient history and examination, biopsies, lab.+ radiological investigations)
- Consider immunosuppressive treatment individually (scarce evidence, patients often multimorbid, side-effects and interactions of medication)
- Other treatment: wound treatment, compression therapy, pain medication, nutrition, psychological support, etc.
- Split-thickness skin-grafting may be considered AFTER the inflammation has been reduced by immunosuppressive therapy
- Multidisciplinary treatment!!
  - Dermatologist often in-charge

Thank you!